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Dermoscopic rainbow pattern: A clue to diagnosing aneurysmal atypical fibroxanthoma

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Abstract: Atypical fibroxanthoma (AFX) is an uncommon spindle cell tumor of fibrous tissue most often located on the sun-damaged skin on the head and neck of elderly patients. It has a locally aggressive behavior and a tendency to recur after surgery but a low metastatic potential.¹ Clinically, AFX presents as a rapidly growing solitary nodule, which sometimes becomes ulcerated.^{2, 3} Several subentities exist, including the aneurysmal atypical fibroxanthoma (AAFX), which is the subject of this report. Dermoscopy of AFX usually shows a nonspecific pattern including slight pigmentation, reddish and whitish areas, and/or a polymorphic vascular pattern (eg, linear, dotted, hairpin, and highly tortuous vessels) irregularly distributed over the surface of the lesion. This pattern can also be seen in other clinically similar tumors, such as squamous cell carcinoma, angiosarcoma, Merkel cell carcinoma, and amelanotic melanoma.^{2, 4, 5} The dermoscopic term rainbow pattern (RP) is used for highly vascularized lesions with areas showing various colors of the rainbow on polarized dermoscopy.⁶ Hemosiderotic and aneurysmal variants of fibrohistiocytic tumors such as dermatofibromas have been described as presenting with a RP.⁷ RP has previously been reported in a case of AFX,⁵ but to the best of our knowledge, no cases of AAFX showing the dermoscopic RP have been reported.

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Dermoscopic rainbow pattern: A clue to diagnosing aneurysmal atypical fibroxanthoma

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Key words: atypical fibroxanthoma; dermoscopy; histopathology; rainbow pattern.

INTRODUCTION

Atypical fibroxanthoma (AFX) is an uncommon spindle cell tumor of fibrous tissue most often located on the sun-damaged skin on the head and neck of elderly patients. It has a locally aggressive behavior and a tendency to recur after surgery but a low metastatic potential.¹ Clinically, AFX presents as a rapidly growing solitary nodule, which sometimes becomes ulcerated.^{2,3} Several subentities exist, including the aneurysmal atypical fibroxanthoma (AAFX), which is the subject of this report.

Dermoscopy of AFX usually shows a nonspecific pattern including slight pigmentation, reddish and whitish areas, and/or a polymorphic vascular pattern (eg, linear, dotted, hairpin, and highly tortuous vessels) irregularly distributed over the surface of the lesion. This pattern can also be seen in other clinically similar tumors, such as squamous cell carcinoma, angiosarcoma, Merkel cell carcinoma, and amelanotic melanoma.^{2,4,5}

The dermoscopic term rainbow pattern (RP) is used for highly vascularized lesions with areas showing various colors of the rainbow on polarized dermoscopy.⁶ Hemosiderotic and aneurysmal variants of fibrohistiocytic tumors such as dermatofibromas have been described as presenting with a RP.⁷ RP has previously been reported in a case of AFX,⁵ but to the best of our knowledge, no cases of AAFX showing the dermoscopic RP have been reported.

Abbreviations used:

AAFX:	aneurysmal atypical fibroxanthoma
AFX:	atypical fibroxanthoma
ERG:	ETS-related gene
RP:	rainbow pattern

CASE REPORTS

Case 1

A 90-year-old male patient presented to the Department of Dermatology at Sahlgrenska University Hospital, Sweden, for a routine skin examination 2 years after surgery for a malignant melanoma. Recently, he had noticed a new skin lesion on the vertex of the scalp. Clinical examination revealed a violaceous nodule measuring 10 × 10 mm surrounded by erythema (Fig 1). A roundish lesion was observed with a diffuse pinkish-red halo and a predominantly violet and white structureless central area. Within the central area, multiple, partly radially arranged, whitish, irregular clods were present. A few of these white clods had a central brown color or were surrounded by violet and red color, producing a RP (Fig 2).

The lesion was excised and histopathologic examination showed dermal atypical spindle cells in an irregular pattern and numerous mitoses. Within the tumor, aneurysmatic spaces filled with red blood cells and hemosiderin were seen. After immunohistochemical examination, angiosarcoma, Kaposi sarcoma, squamous cell

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Fig 1. Violaceous nodule measuring 10 × 10 mm on scalp of 90-year-old male patient with aneurysmal atypical fibroxanthoma (case 1).

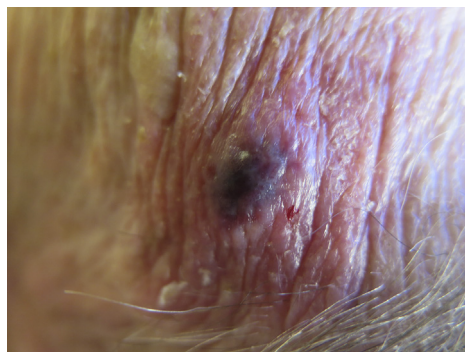


Fig 3. Dome-shaped, bluish-red nodule on the left temple of a 75-year-old male patient with aneurysmal atypical fibroxanthoma (case 2).

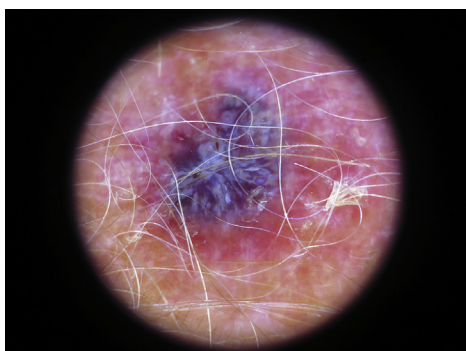


Fig 2. Dermoscopic picture showing a central violet and white structureless area surrounded by a pinkish-red halo. In the center, multiple, partly radially arranged, whitish, irregular clods are present, with a few presenting with a central brown color or surrounded by violet then red color producing a rainbow pattern (case 1).

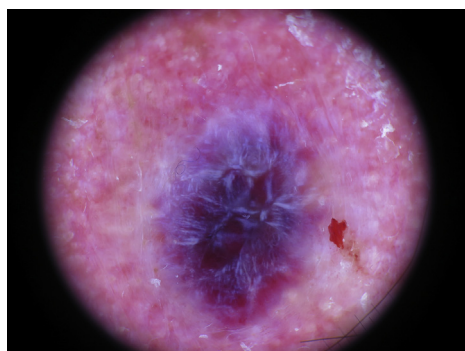


Fig 4. Dermoscopic picture showing a pink-red to dark-blue background color with abundant white shiny lines of varying thicknesses and lengths surrounded by brown, red, and blue colors reminiscent of a rainbow pattern (case 2).

carcinoma, and melanoma were ruled out because CD31, smooth muscle—specific actin, desmin, cytokeratin AE1 and AE3, p40, human melanoma black 45, and melan A were negative. Two pathologists reviewed the case and the diagnosis of AAFX was made.

Case 2

A 75-year-old male patient presented with an intermittently bleeding nodule on his left temple that was first noticed 1 year earlier. His medical history revealed actinic keratoses and a basal cell carcinoma. Clinical examination showed a dome-shaped bluish-red firm nodule with a smooth surface measuring 6 × 10 mm. The raised nodule was clearly demarcated but surrounded by diffuse erythema (Fig 3). Dermoscopically, the lesion elicited a poorly demarcated pink-red to dark-blue background color interspersed with abundant white shiny lines of varying thickness and length, which were partly radially arranged. The white shiny lines

were surrounded by brown, red, and blue colors reminiscent of a RP (Fig 4).

This lesion was excised, and histopathologic examination showed atypical dermal spindle cells, mitoses, bleeding, and surrounding chronic inflammation. The overlying epidermis showed a collision of actinic keratosis with seborrheic keratosis. Immunohistochemical stainings showed high positivity for the proliferation marker Ki67, CD19, and CD99. Some cells showed positivity for CD68 and alpha smooth muscle actin. Stainings negative for desmin, S100, melan A, SOX10, p40, cytokeratin AE1 and AE3, and cytokeratin 20 ruled out leiomyosarcoma, melanoma, squamous cell carcinoma, and Merkel cell carcinoma. Human herpes virus 8, CD31, *ERG* (*ETS*-related gene), and CD34 were absent in stainings of tumor cells and histopathology was not typical for angiosarcoma, Kaposi sarcoma, or dermatofibrosarcoma protuberans. There was, however, positivity for *ERG* and CD31 in some monocytes in the dermal inflammatory infiltrate. Four pathologists reviewed

the case and diagnosed an AFX. The pathologists suggested trauma or inflammation as a possible cause of bleeding.

Because of the recently encountered case of AAFX (case 1) in which dermoscopy revealed an RP on a vascular nodule on the scalp of a patient, the pathologists were asked to review case 2. This histopathologic review led to a final diagnosis of an AAFX rather than AFX with pseudoangiomatous features. The latter diagnosis is another vascular atypical variant of AFX and a histologic and immunohistochemical mimic of cutaneous angiosarcoma.⁸

DISCUSSION

The RP was previously thought to be specific for Kaposi sarcoma but later was described in melanoma, stasis dermatitis, lichen planus, hemosiderotic dermatofibroma, and basal cell carcinoma.^{9,10} Cheng et al revealed that Kaposi sarcoma lesions with the dermoscopic RP histopathologically showed a characteristic vascular lumen-rich structure with back-to-back closely arranged vessels.⁹ Even though different theories exist for the origin of the optical phenomenon of the RP, it seems to be clearly associated with the distinctive microscopic vascular structure of the examined lesions.^{9,10}

This is the first description of the dermoscopic RP in 2 cases of AAFX. In one case, this pattern even led to the correct subclassification of the diagnosis as it helped in the clinicopathologic correlation and gave an explanation for the richness of blood in the tumor. Thus, finding a RP can potentially help dermoscopists shorten their list of differential

diagnoses when assessing amelanotic nodular skin tumors, and subsequently, guide their pathologists in their choice of immunohistochemical stains. In summary, correlation of dermoscopy and histopathology can be key to providing the correct diagnosis of vascular skin tumors such as AAFX.

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